ACC.11
60th Annual Scientific Session & Expo

E229
JACC April 5, 2011
Volume 57, Issue 17

CARDIAC FUNCTION AND HEART FAILURE

THE IMPACT OF SYSTOLIC DYSFUNCTION ON PROGNOSIS OF GENOTYPED HYPERTROPHIC CARDIOMYOPATHY

ACC Poster Contributions
Ernest N. Morial Convention Center, Hall F
Sunday, April 03, 2011, 10:00 a.m.-11:15 a.m.

Session Title: Cardiomyopathies/Myocarditis/Pericardial Disease
Abstract Category: 22. Cardiomyopathies/Myocarditis/Pericardial Disease
Session-Poster Board Number: 1018-34

Authors: *Noboru Fujino, Tetsuo Konno, Akihiko Hodatsu, Kenshi Hayashi, Katsuharu Uchiyama, Takashi Fujita, Toyonobu Tsuda, Hidekazu Ino, Masakazu Yamagishi, Kanazawa University, Kanazawa, Japan*

Background: Hypertrophic cardiomyopathy (HCM) is a disease of sarcomere. Sudden cardiac death in young subjects and heart failure in old subjects are the biggest issues for the management of HCM. Approximately 5% of cases with HCM are reported to show progression to systolic dysfunction (left ventricular ejection fraction (LVEF) <50%), however, few data exist regarding the relationship between the systolic dysfunction and the prognosis in genotyped HCM. Therefore we compared the frequency to progress to systolic dysfunction in genotyped HCM, and assessed the relationship between the systolic dysfunction and the prognosis in genotyped HCM.

Methods: The study comprised of 150 HCM subjects with sarcomere gene mutations. We compared echocardiographic parameters between group-myosin (mutation carriers with cardiac beta-myosin heavy chain gene (n=17) and cardiac myosin binding protein C gene (n=58), mean age=53.5, n=75) and group-troponin (mutation carriers with cardiac troponin T gene (n=22) and cardiac troponin I gene (n=53), mean age=43.0, n=75).

Results: No differences were found in interventricular septal wall thickness (16.1 +/- 5.4 vs 14.6 +/- 5.7 mm), left ventricular end-diastolic dimension (45.5 +/- 6.7 vs. 44.6 +/- 6.5 mm), and LVEF (67.0 +/- 11.9 vs. 65.0 +/- 11.1 %), respectively, between the 2 groups at baseline. Interestingly in the subjects > 40 years of age, the frequency of developing to systolic dysfunction was lower in the myosin-group than in the troponin-group (11.7 vs. 28.3%, P<0.05). Among those who showed systolic dysfunction (LVEF<50%, n=20), we could follow 18 mutation carriers (6 subjects in the myosin-group and 12 subjects in the troponin-group). Seventeen of them experienced admission for heart failure (17/18, 94.4%) and 10 of them died within 10 years after showing systolic dysfunction (10/17, 58.8%).

Conclusions: These results demonstrate that after 40 years of age, mutation carriers in the troponin-group show systolic dysfunction more frequently than those in the myosin-group. Once they demonstrate systolic dysfunction (LVEF<50%), they suffer from heart failure frequently and more than 50% of them die within 10 years.